

Cystic Fibrosis Patients Living Much Longer in 2022 Than in 2008

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Key Findings:

- The median age at death for patients diagnosed with cystic fibrosis (CF) has more than doubled from 26 years of age in 2008 to 66 years of age in 2022.
- Compared to patients without CF, patients with CF have about a 12% lower life expectancy in 2022. This has improved significantly from a 65% lower life expectancy in 2008.
- In 2022, more than 60% of patients diagnosed with CF were 60 years old or older when they died.

Cystic fibrosis (CF) is a disease typically diagnosed in pediatric patients that can have serious impacts on the respiratory, gastrointestinal, endocrine, reproductive, and skeletal systems that limit function and development.¹ In the 1980s and 1990s, CF often led to early death with few patients reaching ages beyond 30 years old. However, advances in treatments and therapies have increased life expectancy for patients both with and without CF throughout the early 2000s and 2010s.¹ We sought to understand how the average age at death for patients with CF compares to patients without CF and whether that has changed over time.

We studied 3,420 patients with CF and 4,783,007 patients without CF who died between 2008 and 2022 to determine the median age at death for each population. We found that the median age at death for patients with CF more than doubled from 26 years in 2008 to 66 years in 2022. The median age at death for patients without CF stayed fairly consistent, with a median of 75 years in both 2008 and 2022. In 2022, the median age at death was just 12% lower for patients with CF compared to patients without CF. This is an 82% reduction of the gap in life expectancy between patients with CF and those without.

Median Age at Death for Patients with and Without Cystic Fibrosis

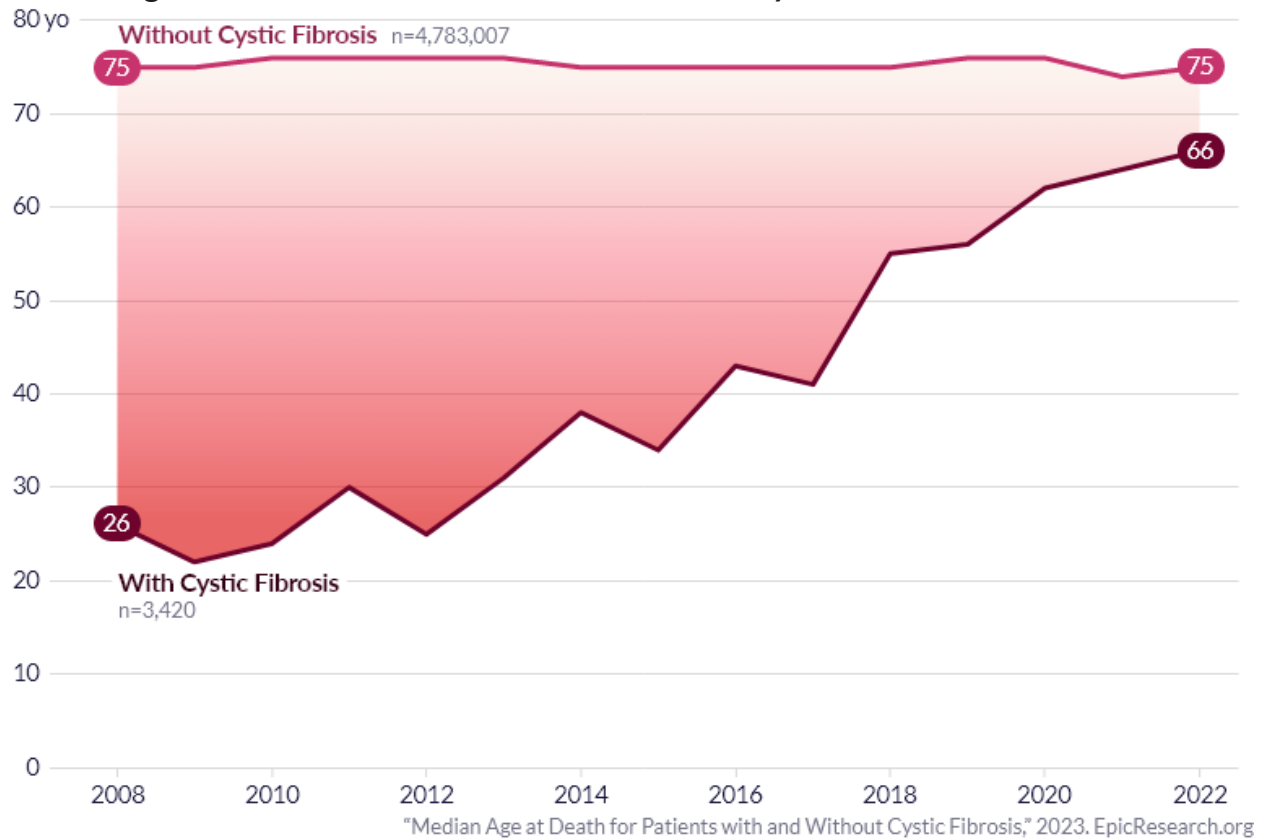


Figure 1. Median age at death for patients who died each year with and without cystic fibrosis from 2008 to 2022.

We further stratified the age at death for patients with CF each year to understand how the proportions of deaths in each decade of life have changed over time. In 2016, nearly half of patients with CF who died were younger than 40. In 2022, that rate was more than cut in half, with about one in five patients with CF who died before age 40.

Proportion of Annual Deaths by Age for Patients with Cystic Fibrosis

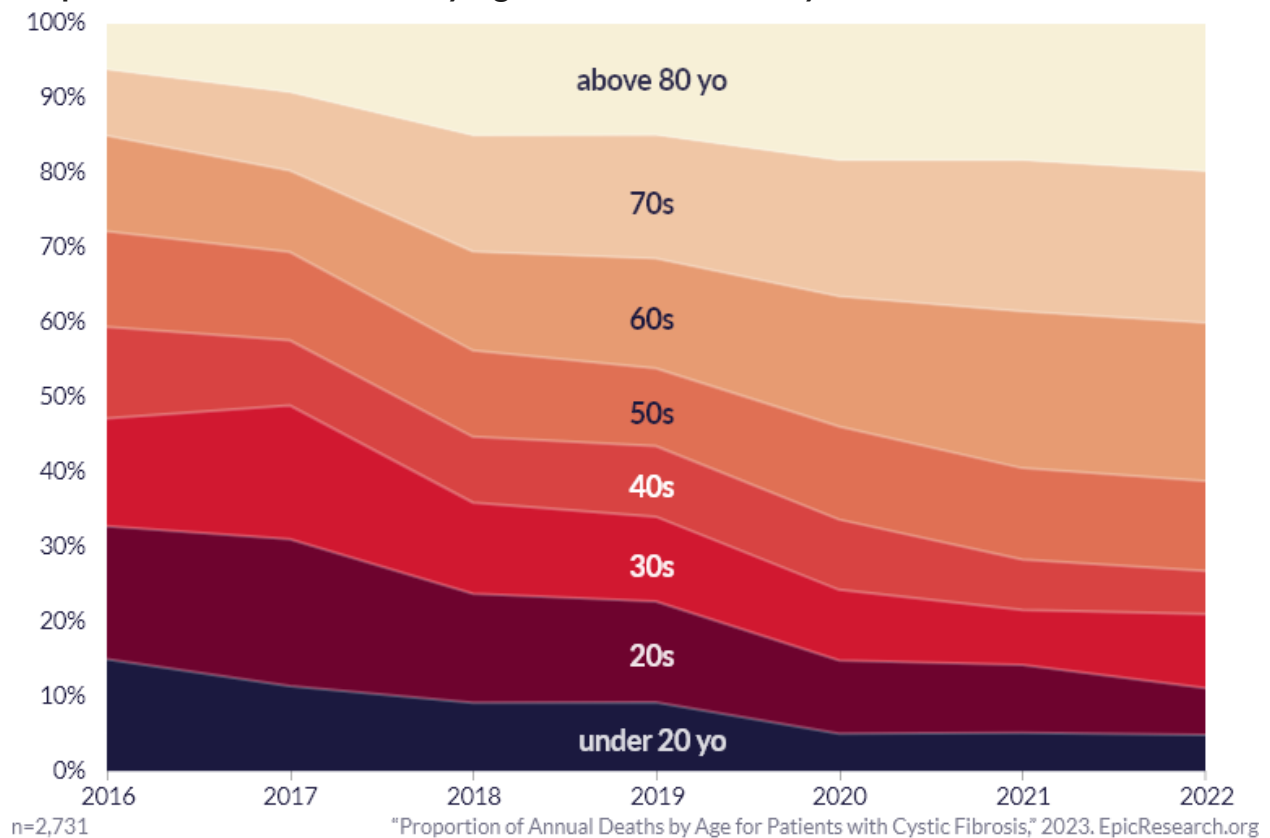


Figure 2. Proportion of deaths for patients with cystic fibrosis in each age group by year of death from 2016 to 2022.

These findings suggest that the advancements in cystic fibrosis treatment and therapies are associated with a substantial increase in life expectancy for patients diagnosed with cystic fibrosis.

These data come from Cosmos, a HIPAA-defined Limited Data Set of more than 203 million patients from 208 Epic organizations including 1,187 hospitals and more than 25,400 clinics, serving patients in all 50 states and Lebanon. This study was completed by two teams that worked independently, each composed of a clinician and research scientists. The two teams came to similar conclusions.

References

1. Link SL, Nayak RP. Review of Rapid Advances in Cystic Fibrosis. *Mo Med.* 2020;117(6):548-554.

Data Definitions

Term	Definition
Study period	2008-2022 (year of death)
Study population	Inclusion criteria: Cystic fibrosis: ICD-10 code: E84.0, E84.11, E84.19, E84.8, or E84.9 Country of residence: USA Death date populated
Exposures	Year of death
Outcomes	Age of death in years

Table 1: Median and Mean Age at Death for Patients with and Without Cystic Fibrosis

Year	Patients with CF deaths	Median age at death - CF (Years)	Mean Age at Death - CF (Years)	95% CI - CF (Years)	Patients without CF deaths	Median age at death - WO CF (Years)	Mean Age at Death - WO CF (Years)	95% CI - WO CF (Years)
2008	33	26	30.33	6.69	45,336	75	70.75	0.18
2009	37	22	26.73	5.88	59,562	75	70.85	0.16
2010	40	24	31.05	5.69	79,865	76	71.48	0.13
2011	56	30	35.39	4.83	113,743	76	71.69	0.11
2012	77	25	30.71	3.99	156,962	76	71.96	0.09
2013	115	31	36.78	3.78	198,428	76	71.99	0.08
2014	162	38	41.83	3.2	233,535	75	72.13	0.07
2015	169	34	40.25	3.5	267,860	75	72.3	0.07
2016	180	43	43.99	3.3	305,340	75	72.15	0.06
2017	229	41	45.72	3.11	390,587	75	72.4	0.06
2018	295	55	51.83	2.73	451,166	75	72.57	0.05
2019	423	56	52.6	2.28	505,154	76	72.86	0.05
2020	499	62	57.92	1.97	621,067	76	72.94	0.04
2021	548	64	59.46	1.88	678,992	74	71.98	0.04
2022	557	66	60.41	1.79	675,410	75	72.68	0.04

Table 2: Proportion of Annual Deaths by Age for Patients with Cystic Fibrosis

Year	<20 years	20-29 years	30-39 years	40-49 years	50-59 years	60-69 years	70-79 years	80+ years
2016	15.0%	17.8%	14.4%	12.2%	12.8%	12.8%	8.9%	6.1%
2017	11.4%	19.7%	17.9%	8.7%	11.8%	10.9%	10.5%	9.2%
2018	9.2%	14.6%	12.2%	8.8%	11.5%	13.2%	15.6%	14.9%
2019	9.2%	13.5%	11.3%	9.5%	10.4%	14.7%	16.5%	14.9%
2020	5.0%	9.8%	9.4%	9.4%	12.4%	17.4%	18.2%	18.2%
2021	5.1%	9.1%	7.3%	6.8%	12.2%	21.0%	20.3%	18.2%
2022	4.8%	6.3%	9.9%	5.7%	12.0%	21.2%	20.3%	19.7%